Intestinal perforation caused by type II enteropathy-associated T-cell lymphoma

Minerva Lazos Ochoa,*‡ Citlali Pasillas Bravo,* Raúl Romero Feregrino,§ Raúl Romero CabelloII

ABSTRACT

Primary lymphomas of the small intestine are rare. Clinical features at presentation and prognosis are variable. T-cell lymphomas may or may not be associated with enteropathy but if it has been previously diagnosed it can present a range of clinical features, show peculiar evolution and affect prognosis. This report describes the case of a patient with insidious gastrointestinal symptoms, who received no treatment, and with no known gastrointestinal alterations. The sudden and unexpected presentation, despite being common with enteropathy-associated T-cell lymphoma (EATL), meant that the diagnosis was unforeseen. It is important to keep in mind the possibility of this diagnosis in the case of a patient with abdominal symptoms and no previous clinical history of this kind of disorder.

INTRODUCTION

Primary lymphomas of the small intestine are rare, accounting for 20-40% of malignant neoplasms found in this site. Clinical features at presentation and prognosis are variable. They can be either of B- or T-cell origin, the former being the most common. T-cell lymphomas may or may not be associated with enteropathy. In most cases, T-cell lymphomas that are not associated with enteropathy present as a high-grade malignancy. In this report, the case of a man with enteropathy-associated T-cell lymphoma (EATL II) is presented whose major clinical manifestation was intestinal perforation.

CLINICAL SUMMARY

A 46-year-old man with no significant medical antecedents presented with evidence of intestinal obstruction, lower gastrointestinal bleeding, weight loss, and suffering from septic and hypovolemic shock. He died with these conditions short after his arrival at the emergency room.

AUTOPSY SUMMARY

An ileal perforation was found 15 cm from the ileocecal valve. It was 5 cm along its major axis but 95% sealed by the greater omentum. There was an associated transmural neoplastic lesion, which had a well-defined prominence towards the luminal surface, which was ulcerated and 7.3 cm along its major axis (figure 1). Histologically, the lesion had the characteristics of a lymphoid neoplasm with a diffuse growth pattern and infiltrating edges up to the muscle layer (figure 2). It was composed of round-to-polygonal cells of medium-to-large size with scant cytoplasm and large central pleomorphic nuclei, some with vesicular chromatin and prominent nucleoli (figure 3). The adjacent mucosa showed villous atrophy (figure 4). The neoplastic cells were CD3, CD8 and CD56

Key words:

Palabras clave:
Enteropatía asociada a linfoma de células T, linfomas de células T, enteropatía.
positive by immunohistochemical staining (figure 5). An increased number of CD4 and CD8 positive intraepithelial lymphocytes (IEL) were detected in the mucosa adjacent to the lesion (figure 4), as well as fibroadhesive peritonitis localized to the area neighboring the perforation. The rest of the organs showed no alterations. Based on these findings a diagnosis of EATL was made.

**DISCUSSION**

EATL was first described as a neoplasm associated with celiac disease in 1978 by Isaacson and Wright, and Isaacson and collaborators demonstrated its T-cell origin in 1986. It was O’Farrell and colleagues, however, who coined the term EATL because of the close association of the lesion with villous atrophy in the jejunal mucosa adjacent to the disease. At the lymphoma workshop of the XVIth meeting of the European Association for Haematopathology and the Society for Hematopathology was proposed that the EATL could be of two types EATL I and EATL II each one with clearcut differences (table I): the type I has evidence of coeliac disease, usually is alfa-beta, often double-negative for CD4 and CD8, and polymorphous cytological composition.9 While type II also known as (monomorphic intestinal T-cell lymphoma) usually has not evidence of coeliac disease, has a worldwide distribution, usually gamma-delta, monomorphic, and CD8 and CD56 positive.10-12

Primary lymphomas of the small intestine have a different clinical presentation and prognosis, and differ-
The lesions may be single or multiple, usually in the form of circumferential small bowel ulcers. Lesions can also be nodules, plaques or stenotic, but rarely appear as big masses. The mesenteric lymph nodes are often enlarged by infiltration or by reactivity to the disease. The lesions widened the villi, and is comprised of small-to-medium cubic cells, with scant eosinophilic cytoplasm and large nuclei, in which hyperchromatic nucleoli can be observed. The presence of mitotic figures and villous atrophy is frequent, as well as crypt hyperplasia in the mucosa adjacent to the lesion.

The diagnosis is almost always made with a laparotomy. Immunoproliferative small bowel disease and coeliac disease should be considered in the differential diagnosis, especially in cases where villous atrophy as well as symptoms predominantly of malabsorption are evident.

Patients are typically treated with a combination of surgery, to remove as much of the tumor as possible, and chemotherapy. Staging is carried out by Axial Computer Tomography (ACT) and a bone marrow biopsy.

**COMMENT**

The frequency of intestinal T-cell lymphoma is approximately 30%, and it is associated with celiac disease in 5% of cases. There is, however, a rare variant where patients have no previous symptoms and which shows characteristic histopathological features, namely, monomorphic cells, increased CD8 and CD56 positivity (80-90%) compared to intestinal T-cell lymphoma associated with celiac disease, and c-MYC amplification. These factors should be considered in the diagnosis of such neoplasms.

The prognosis of patients with EATL is poor, with a 2 year overall survival rate of approximately 28%. There are no reports on the frequency of this neoplasm in Mexico.

This report describes the case of a patient with insidious gastrointestinal symptoms, who received no treatment, and with no known gastrointestinal alterations. The sudden and unexpected presentation, despite being common with EATL, meant that the diagnosis was unforeseen. It is important to keep in mind the possibility in over two thirds of cases, often accompanied by a 25% increased level of LDH. Serum albumin is usually diminished. Less than half of cases with celiac disease and lymphoma have the two conditions diagnosed at the same time.

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Type I enteropathy-associated T-cell lymphoma</th>
<th>Type II enteropathy-associated T-cell lymphoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Frequency</td>
<td>80-90%</td>
<td>10-20%</td>
</tr>
<tr>
<td>Morphology</td>
<td>Variable (large pleomorphic cells)</td>
<td>Small monomorphic cells</td>
</tr>
<tr>
<td>Immunophenotype</td>
<td>CD3 Positive</td>
<td>Positive</td>
</tr>
<tr>
<td></td>
<td>CD4 Negative</td>
<td>Negative</td>
</tr>
<tr>
<td></td>
<td>CD8 Negative to 20% positive</td>
<td>Positive</td>
</tr>
<tr>
<td></td>
<td>CD56 Negative</td>
<td>Positive to 30-40% positive</td>
</tr>
<tr>
<td></td>
<td>HLA-DQ2/DQ8 90% positive</td>
<td>Villous atrophy</td>
</tr>
<tr>
<td></td>
<td>Mucosa Villous atrophy</td>
<td>Increase in CD8+ lymphocytes</td>
</tr>
<tr>
<td>Genetics</td>
<td>+8q24 (MYC) 27%</td>
<td>73%</td>
</tr>
<tr>
<td>TCR rearrangement</td>
<td>Present</td>
<td>Present</td>
</tr>
<tr>
<td>EBV</td>
<td>Negative</td>
<td>Negative</td>
</tr>
</tbody>
</table>

TCR = T-cell receptor, EBV = Epstein-Barr virus.
of this diagnosis in the case of a patient with abdominal symptoms and no previous clinical history of this kind of disorder.

REFERENCES